CASE REPORT

Recurrent Calcifying Fibrous Tumour in Axilla: A Rare Case Report

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INTRODUCTION

Calcifying fibrous tumour is a rare benign lesion. Calcifying fibrous tumor (CFT), synonymous with a calcifying fibrous pseudotumor, is a rare, distinct tumor-like soft tissue lesion that usually affects children and young adults, with in their subcutaneous and deeps of tissues of extremities, trunk, neck and scrotum[1,2].

Calcifying fibrous tumor (CFT), originally defined by Rosenthal and Abdul-Karim (1) as a childhood fibrous tumor with psammoma bodies, was subsequently renamed by Fetsch et al(2)as a calcifying fibrous pseudo tumor. It has tendency of non destructive local recurrence. Histologically, it is characterised by a non-encapsulated, well-circumscribed mass, containing dense hyalinised collagen, bland spindle cells, lymphoplasmacytic infiltrate sand psammoma bodies, or diastrophic calcifications.(3)

CASE REPORT

A 7 year-old child presented with slow growing swelling over left axilla. Patient was operated for similar swelling 4 years back. Clinical examination revealed a firm, well defined mobile non tender swelling of size 8x5 cm in left axilla. The patient's complete blood count and other routine bio-chemical values were within normal limits. There was no family history. MRI scan revealed multiple altered signal intensity masses in left axillary region, few of them are conglomerated to each other with internal hypo intensities. Excisional biopsy was performed and sent for histopathological examination.

Grossly three grey white to grey brown well circumscribed, lobulated firm soft tissue masses of size 8x8x3.5, second 6.5x4.5x3.5 and smallest was of 2x1.5x1cm were present. External surface of all masses was capsulated globular grey white. Cut surface of all was fibrous grey white.

Microscopically sections show presence of loose hyalinized bands of collagen, hypocellular in nature, along with areas of dystrophic calcification,presence of lymphoid follicles with germinal centres and large number of reactive plasma cells favouring calcifying fibrous pseudotumour.

DISCUSSION

Calcifying fibrous tumor (CFT) is a rarely seen entity that usually occurs in children and young adults, with a slight increase in risk in women. Clinically, it appears as a slow growing, non tender mass.

These lesions were originally described as arising in the sub cutaneous and deep soft tissues, mostly in the extremities and the neck areas[2] but more recently CFT have been found to be ubiquitous.

Most lesions are solitary but multi focal lesions are also reported specially arising from pleura and peritoneum and uncommonly in mesentry[4,5]. Malignant transformation has not been reported[6].

A few were thought to evolve from inflammatory myofibroblastic tumor(IMT), but larger studies failed to confirm this and ultra structural studies revealed fibroblastic features[4,7]. However, in the literature, there are cases that maybe related to trauma and Castleman's disease[8-12].

Nascimen to et al have observed local recurrences in 3 of 10 patients in his study[13]. Ultra structural studies show that calcifications occur as a result of cytoplasmic degeneration in the fibroblasts[10,13]. The local recurrence rate for CFT has been estimated to be approximately 10%. the rate of recurrence in gastrointestinal CFTs as opposed to CFTs arising at other sites remains unclear. Some authors have suggested that gastric CFTs, for example, have no tendency for local recurrence compared to soft tissue CFTs[14].

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The his to pathological features of CFTs are generally easily recognizable from other reactive or benignneo plastic lesions. Inflammatory myofibroblastic tumor (IMT), Plexi form fibromyxoma, reactive nodular fibrous pseudo tumor(RNFP), nodularfasciitis, desmoidfibromatosis, giant cell fibroma, desmoplastic fibroblastoma, fibroma of tendon sheathand calcifying aponeurotic fibroma might be considered in the differential diagnosis. Amyloid fibromas show positive staining with Congo red[15].

There are few studies regarding cytogenetic abnormalities in calcifying fibrous tumors. By using fluorescent in situhybridization, Hoffmann et al attempted to detect trisomy 7 and trisomy 8 which has been reported in other benign fibrous tumors. Due to the low signal intensity, they were not able to complete the study[16]

In another study, Fukunaga et al found that CFT had a diploid DNA contentby flow cytometry[17]. Whether CFT is a true neoplasm or are active process still remains unknown[16-18]. Future molecular studies can be helpful for detecting the biologicbehavior of CFT.

Further studies are needed to clarify the recurrence rate of CFTs in different anatomic sites. Such studies could drive guidelines for management of patients with CFT. In summary, CFT is a rare soft tissue neoplasms and conservative excision is sufficient to cure the patient. However, local recurrence can be seen in some cases. Thus clinical follow up is important. These lesions may be identified more often in the future due to increased awareness and technical advances in diagnosis.

Fig 1: Gross Specimen

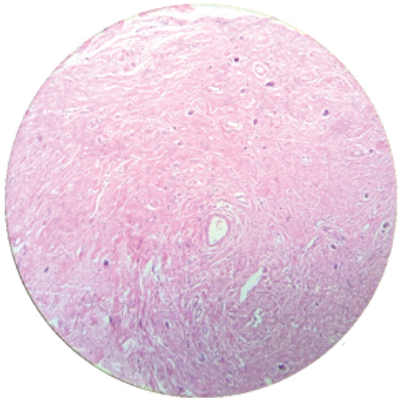

Fig 2: 4x

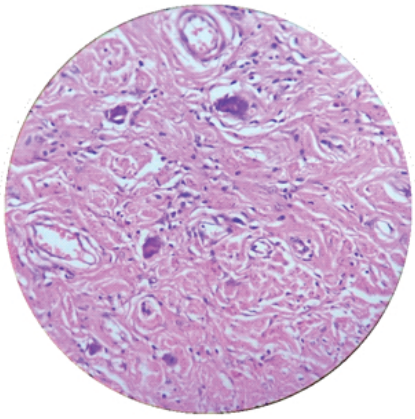

Fig 3: 40x

REFERENCES:

1. Rosenthal NS, Abdul-Karim FW. Childhood fibrous tumor withpsammoma bodies. Clinicopathologic features in two cases. ArchPatholLabMed1988;112:798-800.
2. FetschJF,MontgomeryEA,MeisJM. Calcifyingfibrouspseudotumor.AmJSurgPathol1993;17:502-8.
3. Diagnostic histopathology of tumors volume 2 4th edition. Christopher Fletcher.
4. NascimentoAF,RuizR,HornickJL, FletcherCD.Calcifyingfibrous "pseudotumor": clinicopathologicstudyof15casesandanalysisofitsrelationshiptoinflammatory myofibroblastictumor. *IntJSurgPathol*.2002;10:189e196.
5. ChenKTK.Familialperitoneal multifocalcalcifyingfibroustumor.*AmJClinPathol*. 2003;119:811e815.
6. Farah RB,Dimet S, BidaultAT,et al. Multiple peritonealcalcifyingfibroustumorsrevealedbyischemiccolitis.*AnnDiagnPathol*.2007;11:460e463.
7. Sigel JE, Smith TA, Reith JD, Goldblum JR.Immunohistochemical analysis of anaplastic lymphomakinaseexpressionindeepsofttissuecalcifyingpseudotumor:evidenceofalatesclerosingstageofinflammatorymyofibroblastictumor?*AnnDiagnPathol*.2001;5:10e14.
8. BellDM,DekmezianRH,HusainSA, LunaMA. Oralcalcifyingfibrouspseudotumor:Caseanalysisandreview.HeadNeckPathol.2008;2:343-7.
9. Azam M, Husen YA, Pervez S. Calcifying fibrous pseudotumor inassociation with hyalinevasculartype Castleman' sdisease. IndianJPatholMicrobiol. 2009;52:527-9.
10. ZamecnikM, DorociakF, VeselyL. Calcifyingfibrouspseudotumoraftertrauma. PatholInt.1997;47:812
11. PomplunS,GoldstrawP, DaviesSE, BurkeMM, NicholsonAG. Calcifying fibrouspseudotumourarising with inaninflammatory pseudotumour: Evidence of progression from one lesion to theother? Histopathology.2000;37:380-2.
12. HillKA,Gonzalez-CrussiF, ChouPM. Calcifyingfibrouspseudotumorversusinflammatory myofibroblastictumor:Ahistological and immunohistochemical comparison. Mod Pathol.2001;14:784-90.
13. Nascimento AF, Ruiz R, Hornick JL, Fletcher CD. Calcifyingfibrous 'pseudotumor': Clinicopathologic study of 15 cases andanalysisofitsrelationshiptoinflammatorymyofibroblastictumor.IntJSurgPathol.2002;10:189-96.
14. **George SA**, Abdeen S. Gastric Calcifying Fibrous Tumor Resembling Gastrointestinal Stromal Tumor: A Case Report. *Iran J Pathol* 2015; **10**: 306-309 [PMID: 26351502]
15. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Weiss SW, GoldblumJR,editors. Mosby Elsevier;2008.294-7
16. HoffmannH,BeaverME,MaillardAAJ. Calcifyingfibrouspseudotumoroftheneck.ArchPatholLabMed.2000;124:435-7.
17. Fukunaga M, Kikuchi Y, Endo Y, Ushigome S. Calcifying fibrouspseudotumor. PatholInt. 1997;47:60-3.
18. Jiang K, Nie J, Wang J, Li J. Multiple calcifying fibrous pseudotumorofthebilateralpleura. JpnJClinOncol.2011;41:130-3.